2:30

794-3 Development and Validation of a Conductance Method to Measure Pulmonary Regurgitation in Patients Late After Tetralogy of Fallot Repair

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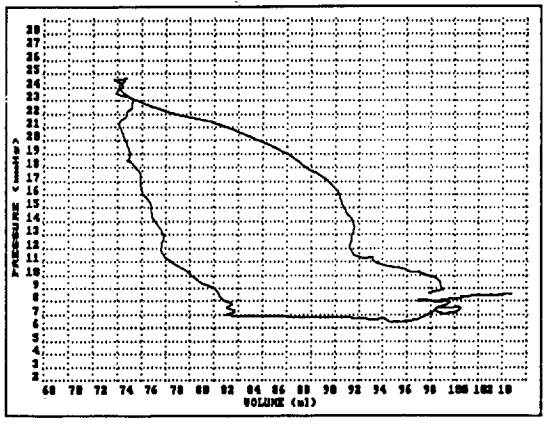
Pulmonary regurgitation (PR) is a significant determinant of right ventricular and global functional performance late after repair of tetralogy of Fallot (rTOF). We have previously shown that PR can be quantified using pressure volume loops by measuring RV volume increase during isovolumic relaxation period. This (digitisation of RV angiograms) and other methods of RV volume measurement (eg. MRI) do not allow real time analysis. However, measurement of RV blood volume by a conductance catheter may allow beat to beat analysis of RV volume and PR fraction. A conductance catheter method was validated which would enable continuous on line recording of pressure volume loops from which pulmonary incompetence could be directly assessed. Five silicone human right ventricular casts were constructed and an assessment of the accuracy and linearity of the conductance catheter was undertaken to compare conductance against known volume. The mean correlation coefficient (r^2) and Y intercept for all the right ventricular cast studies was 0.94 (SE 0.02) and −0.225 (SE 1.72) respectively. Five patients late after rTOF with clinically significant PR had right heart catheterisation during which simultaneous microtip pressure and conductance volume were measured. All five patients had pressure volume loops typical of PR (Figure). The PR volume was indexed to stroke volume and varied significantly with respiration ($p < 0.05$), decreased with Valsalva manoeuvre ($p < 0.05$) and increased with transient right pulmonary artery occlusion ($p < 0.05$). Conductance measurement allows real time dynamic assessment of PR and demonstrates the impact of different loading conditions.



2:45

794-4 Compensated Pulmonic Insufficiency: Is the Right Ventricular Response to Exercise Normal?

Philip J. Iuliano, Stephen E. Cyran, William R. Davidson, Jr.. *Penn State University, Hershey, PA*

Chronic right ventricular (RV) volume overload due to pulmonic insufficiency (PI) can unpredictably lead to irreversible RV failure. We sought to assess the effect of chronic volume overload on RV exercise response. 10 patients (2 male, 8 female, mean age 24 yrs) with compensated moderate to severe PI due to surgically corrected Tetralogy of Fallot or pulmonic stenosis and 7 controls (4 male, 3 female, mean age 29 yrs) were studied on a symptom limited stationary cycle progressive ramp protocol with measurement of VO_2 and respiratory quotient. Stress echocardiography was performed at rest and during exercise (mid = RQ 0.85, 100% = peak). RV function was described as end diastolic RV area (RVEDA), end diastolic RV/LV area ratio (RV/LV) and percent systolic RV area change (%RVAΔ), the latter a reflection of RV ejection fraction.

Results:

	patients (P)			controls (C)		
	rest	mid	100%	rest	mid	100%
RVEDA*	28.5	26.4	24.7	19.2	16.1	13.8
RV/LV**	0.98	0.95	0.97	0.56	0.53	0.49
%RVAΔ†	0.37	0.36	0.34	0.34	0.41	0.43

By ANOVA P vs C *$p < 0.005$, **$p < 0.001$

Although there was no significant difference in percent systolic RV area change at rest, in patients RV systolic function declined during exercise as compared with an increase in controls, †$p < 0.001$. Cardiac index, HR, and BP did not significantly differ between patients and controls. VO_2 max was reduced in patients with PI (23.0 vs 38.8 ml/kg/min, $p < 0.001$), but was not predicted by any resting echocardiographic variable. However, in controls larger RVEDA was associated with a greater exercise capacity. The change in RV systolic performance during exercise was associated with resting RVEDA, RV/LV and %RVAΔ, $p < 0.005$.

Conclusion: Patients with compensated significant PI have an abnormal RV response to exercise demonstrable by stress echocardiography. This may have implications for the timing of pulmonic valve replacement.

3:00

794-5 Long-term Survival in Unoperated Single Ventricle

Naser M. Ammash, Carole A. Warnes *Mayo Clinic, Rochester, Minnesota*

Survival into adulthood is rare in patients with single ventricle without palliative surgery or radical repair. The Fontan operation is commonly utilized in those patients with ideal hemodynamics in the hope of conferring improved survival.

From 1984–1994, 12 patients (7 males, 5 females) aged 23–66 years (mean 41) with a single ventricle and no prior surgery were evaluated. Eleven had double-inlet left ventricle (DILV), one had tricuspid atresia, and of these 12, 8 had moderate to severe pulmonary stenosis with a peak gradient from 71–112 mmHg (mean 94). The other four had moderate to severe pulmonary hypertension (PH) with a pulmonary artery systolic pressure ranging from 78–122 mm Hg (mean 102). Six of these 12 patients were followed from 124 years (mean 14 yrs). Eleven patients had an ability index of 2, and one had an ability index of 3. Plasma hemoglobin ranged from 15.3 – 21.8 gm/dL and was higher in those with PH. Only 4 of 12 patients had had phlebotomy. Ventricular function by 2-D echocardiogram or left ventriculography was normal in 10 patients and mildly depressed in 2 patients. Two of the 12 patients were on diuretics. Rhythm was sinus in 10 patients; two others had a history of atrial fibrillation controlled on medication. Two patients had a permanent pacemaker, one for chronic atrial fibrillation with slow ventricular response and the other for complete heart block. One 60-year-old patient with a history of stroke died from a second stroke.

While this group is biased by referral and natural selection, patients demonstrated excellent functional capacity and preserved ventricular function. Adult patients with DILV and perfectly balanced pulmonary circulation may do as well as, if not better than, those with a Fontan operation who have a 60% 10-year survival. This should be considered before sending an adult patient with single ventricle for a Fontan repair.

3:15

794-6 Functional Capacity in Adult Patients with Mustard Correction

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The effect of prolonged support of the systemic circulation by the right ventricle was assessed in adult survivors of the Mustard operation. Forty five adult patients (32 male) aged 21.7 +/− 3.3 years (mean +/− S.D) were studied. They had undergone Mustard's operation for correction of complete transposition 19.1 +/− 2.6 years previously at 2.6 +/− 3.1 years of age. Cardiopulmonary function was assessed using maximal bicycle ergometry with respiratory mass spectrometry. Systemic ventricular ejection fraction (SVEF) was measured at rest (r) and with exercise (ex) by radionuclide angiography. Thirty four patients additionally had magnetic resonance imaging to measure ventricular dimensions. Results were compared with normals [mean (SD)]

	Max. Work load kpm	VO2 max	AT	SVEF (r)	SVEF (ex)	SVTh	SVD	% NYHA 1 or 2
Mustard	673*	17.1*	11.5*	45.4*	51.1*	14.0*	40.4+	92
	(179)	(4.9)	(4.0)	(10.9)	(14.7)	(3.7)	(7.4)	
Normal	1117	43.4	24.2	59.9	70.6	9.3	32.5	100
	(190)	(10.1)	(4.7)	(8.7)	(7.7)	(1.0)	(3.5)	

AT = anaerobic threshold, VO2 max = maximal oxygen consumption, SVD = systemic ventricular diameter (mm). SVTh = wall thickness (mm); *$p < 0.01$, +$p < 0.05$

There was no significant difference in maximal heart rate or maximal blood pressure with exercise between the groups. There was no correlation between ventricular dimension/thickness and ejection fraction either at rest or exercise. Maximal work load was correlated with ejection fraction on exercise ($p < 0.05$)

Thus adult Mustard patients have dilated, hypertrophied systemic ventricles with reduced ejection fraction compared with controls. Formal functional assessment demonstrates significant impairment despite patients leading normal or near normal lives.